

Claims

1. Use of a peptide or a protein selected from Apolipoprotein B; a fragment thereof; Apolipoprotein E and a fragment thereof, in an assay for the detection of the formation of PrP^{Sc} in a sample.
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2. Use of a peptide or a protein selected from Apolipoprotein B; a fragment thereof; Apolipoprotein E and a fragment thereof, in a screening assay for identifying compounds that modulate the conversion of PrP^C into PrP^{Sc}.
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3. Use according to claims 1 or 2 wherein the peptide or protein is selected from Apolipoprotein B; a fragment thereof; Apolipoprotein E and a fragment thereof, which forms a complex with the LDL receptor.
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4. Use according to any of the preceding claims wherein the assay is a Protein Misfolding Cyclic Amplification (PMCA) assay.
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5. Use according to any of the preceding claims wherein the assay is a Protein Misfolding Cyclic Amplification (PMCA) assay using normal brain homogenate 20 as a source of normal PrP^C and substrate.
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6. Use according to claims 1 to 4 wherein the assay is a Protein Misfolding Cyclic Amplification (PMCA) assay using lipid rafts from infection sensitive neuroblasma cell line N2a as a source of normal PrP^C and substrate.
7. Use according to any of the preceding claims wherein the protein is Apolipoprotein B.
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8. Use according to any of the preceding claims wherein the protein is Apolipoprotein B, the assay is a Protein Misfolding Cyclic Amplification (PMCA) assay using lipid rafts from infection sensitive neuroblasma cell line N2a as a source of normal PrP^C and substrate.

9. Use of a modulator of a protein or a peptide, wherein the protein is selected from from Apolipoprotein B and a fragment thereof, for the preparation of a pharmaceutical composition for the treatment of a prion disease.

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10. Use according to claim 9 wherein the modulator is an antibody raised against Apolipoprotein B or a fragment thereof.

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11. Use according to any of the preceding claims wherein the peptide or the protein contains the sequence of SEQ ID NO: 3.

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12. Use according to any of the preceding claims wherein the peptide or the protein is of a molecular weight selected from 30 and 40 kDa and which sequence is selected from fragments of Apolipoprotein B taken between positions 3201-3558, 3548-3905, 3201-3905, 3291-3558, 3548-3815 and 3291-3815.

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13. Use according to any one of claims 9 to 12 wherein the prion disease is bovine spongiform encephalopathy (BSE).

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14. Use according to any one of claims 9 to 12 wherein the prion disease is a Creutzfeld-Jacob Disease (CJD).

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15. A method for the diagnosis or detection of a prion disease within a subject suspected of suffering from such a disease which comprises (i) contacting a sample from said subject with a peptide or a protein selected from Apolipoprotein B; a fragment thereof; Apolipoprotein E and a fragment thereof; (ii) contacting the mixture obtained in step (i) with PrP^C or PrP^C containing mixtures; and (iii) determining the presence and/or amount of PrP^{Sc} in said sample.

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16. A method of determining a marker that predisposes a subject to a prion disease, comprising (i) measuring a level of a protein selected from Apolipoprotein B; a

fragment thereof; in said sample; and (ii) correlating said level of protein obtained in said measuring step with the occurrence of a prion disease.

17. A method according to any one of claims 15 to 16 wherein the prion disease is
5 bovine spongiform encephalopathy (BSE).

18. A method according to any one of claims 15 to 16 wherein the prion disease is a
Creutzfeld-Jacob disease.

10 19. A method for the detection of PrP^{Sc} within a sample, which assay comprises (i) contacting said sample with a peptide or a protein selected from Apolipoprotein B; a fragment thereof; Apolipoprotein E and a fragment thereof; (ii) contacting sample obtained in (i) with PrP^C or PrP^C containing mixtures; and (iii) determining the presence and/or amount of PrP^{Sc} in said sample.

15 20. A method for identifying, in a sample, a compound which modulates the transition of PrP^C into PrP^{Sc} comprising: (i) contacting said sample with a peptide or a protein selected from Apolipoprotein B; a fragment thereof; Apolipoprotein E and a fragment thereof; (a) in the presence of said modulatory compound and (b) in the absence of said compound; (ii) contacting the mixtures obtained in step (i) a and (i) b with PrP^C or PrP^C containing mixtures; and (iii) determining the amount of PrP^{Sc} (a) in the presence of said modulatory compound and (b) in the absence of said modulatory compound.

25 21. A method according to any one of claims 15 to 20 wherein the peptide or the protein contains the sequence of SEQ ID NO: 3.

30 22. A method according to any one of claims 15 to 21 wherein the peptide or the protein is of a molecular weight selected from 30 and 40 kDa and which sequence is selected from fragments of Apolipoprotein B taken between positions 3201-3558, 3548-3905, 3201-3905, 3291-3558, 3548-3815 and 3291-3815.

23. An assay for the detection of PrP^{Sc} in a sample, which assay comprises (i) contacting said sample with a peptide or a protein selected from Apolipoprotein B; a fragment thereof; Apolipoprotein E and a fragment thereof; (ii) contacting the mixture obtained in step (i) with PrP^C or PrP^C containing mixtures; (iii) determining the presence and/or amount of PrP^{Sc} in said sample.

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24. A screening assay for identifying a compound which modulates the transition of PrP^C into PrP^{Sc} comprising: (i) contacting said sample with a peptide or a protein selected from Apolipoprotein B; a fragment thereof; Apolipoprotein E and a fragment thereof; (a) in the presence of said modulatory compound and (b) in the absence of said modulatory compound; (ii) contacting the mixtures obtained in step (i) a and (i) b with PrP^C or PrP^C containing mixtures; and (iii) determining the amount of PrP^{Sc} (a) in the presence of said compound and (b) in the absence of said modulatory compound.

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25. An assay according to any one of claims 23 to 24 wherein the peptide or the protein contains the sequence of SEQ ID NO: 3.

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26. An assay according to any one of claims 23 to 25 wherein the peptide or the protein is of a molecular weight selected from 30 and 40 kDa and which sequence is selected from fragments of Apolipoprotein B taken between positions 3201-3558, 3548-3905, 3201-3905, 3291-3558, 3548-3815 and 3291-3815.

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27. A diagnostic kit for use in an assay according to claims 23 to 26, comprising a probe for receiving a sample and a peptide or a protein selected from Apolipoprotein B and a fragment thereof.

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28. An apparatus for use in a method according to any one of claims 15 to 22 or an assay according to any one claims 23 to 26.